Cardiology in the Young (2017), **27**, 1637–1640
doi:10.1017/S1047951117000798

Brief Report

Flail anterior tricuspid valve leaflet in a neonate: association with maternal antiphospholipid syndrome

Adrian J. Tarca,[1] Luke Eckersley,[1,2] Darshan Kothari[1]

[1]*Children's Cardiac Centre, Princess Margaret Hospital;* [2]*University of Western Australia, Perth, Western Australia, Australia*

Abstract This brief report describes a case of flail anterior tricuspid valve leaflet in a neonate associated with maternal antiphospholipid syndrome. Fetal echocardiography at 27 weeks of gestation showed competent atrioventricular valves with biventricular echogenic chordae. Fetal distress was noted at delivery, and echocardiography showed a flail anterior leaflet of the tricuspid valve with severe regurgitation. Possible causation and implications of maternal antiphospholipid syndrome are discussed.

Keywords: Tricuspid regurgitation; flail leaflet; maternal antiphospholipid syndrome; echogenic chordae

Received: 26 November 2016; Accepted: 20 March 2017; First published online: 23 May 2017

Tricuspid regurgitation due to flail anterior leaflet secondary to chordae rupture is a rare, life-threatening event.[1–3] This case is of interest because of the possible involvement of maternal antiphospholipid syndrome, not previously reported in the literature.

The patient was conceived via gamete intrafallopian transfer by a G_2P_0 mother aged 31 years. There was a maternal history of antiphospholipid syndrome, with positive anti-SSA/Ro and antiphospholipid antibodies, and previous spontaneous abortion at 16 weeks. She was managed with aspirin, enoxaparin, and vitamin D.

Routine ultrasound at 19 weeks of gestation showed an abnormal tricuspid valve appearance, prompting cardiology review. Fetal echocardiogram at 21 weeks of gestation noted echogenic chordae in both the right and the left ventricle, but with competent atrioventricular valves (Fig 1). Repeat echocardiogram at 27 weeks of gestation was unchanged, and routine obstetric/neonatal management was advised. (Supplementary videos 1 and 2).

A female infant was born at 38 + 6 weeks of gestation by emergency caesarean section because of suspected fetal compromise. Her birth weight was 3230 g, and her Apgar scores were 7^1, 9^5. Oxygen resuscitation was required because of desaturation to 65%. The patient was transferred to the tertiary neonatal unit for ongoing care, with cardiology consultation.

On examination, the patient was cyanosed, and a 3/6 pan-systolic murmur was audible, loudest at the lower left sternal edge, radiating throughout the chest. There was no diastolic component, and she had normal peripheral and femoral pulses. Chest radiography showed cardiomegaly. No clinical or investigational evidence for neonatal lupus was observed, including rash, heart block, cardiomyopathy, or prolonged QTc; however, the bright papillary muscles may represent endocardial fibroelastosis.

Subsequent echocardiography revealed a flail anterior leaflet of the tricuspid valve and severe tricuspid regurgitation (Fig 2). The right heart was dilated, with good right ventricular function. Mildly bright right ventricular papillary muscles were noted. A patent ductus arteriosus showed bidirectional flow, and the patent foramen ovale flow was right to left. Owing to severe tricuspid regurgitation, there was functional pulmonary atresia. In addition, there was circuit reversal, with pulmonary regurgitation observed in association with ductal flow. Cardiac connections and structure were otherwise normal (Supplementary video 3).

Correspondence to: Dr A. J. Tarca, Paediatric Resident, Children's Cardiac Centre, Princess Margaret Hospital, Roberts Road, Subiaco, Perth, WA 6008, Australia. Tel: 08 9340 8222; Fax: 93408983; E-mail: Adrian.Tarca@health.wa.gov.au

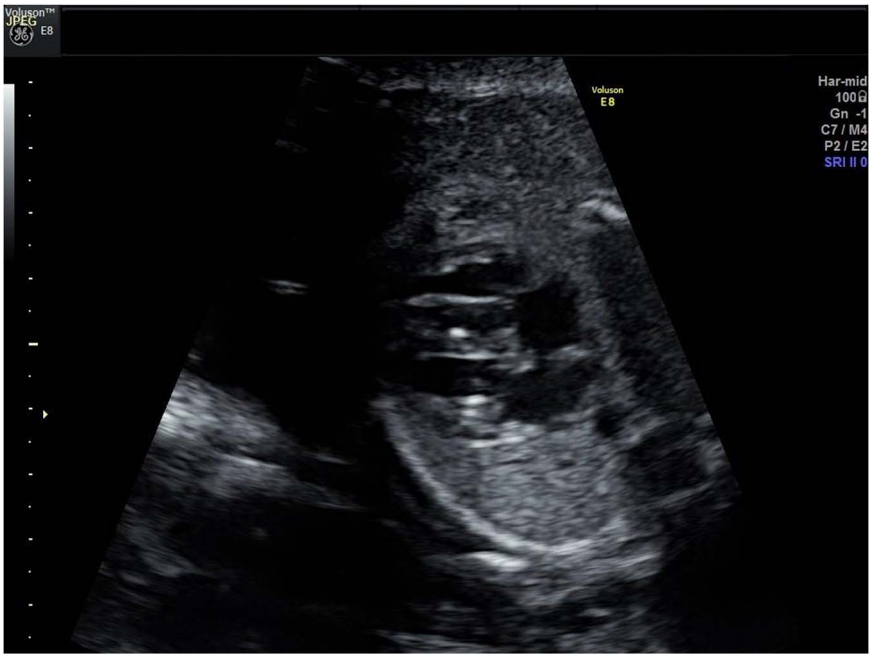

Figure 1.
Fetal echocardiogram showing echogenic chordae.

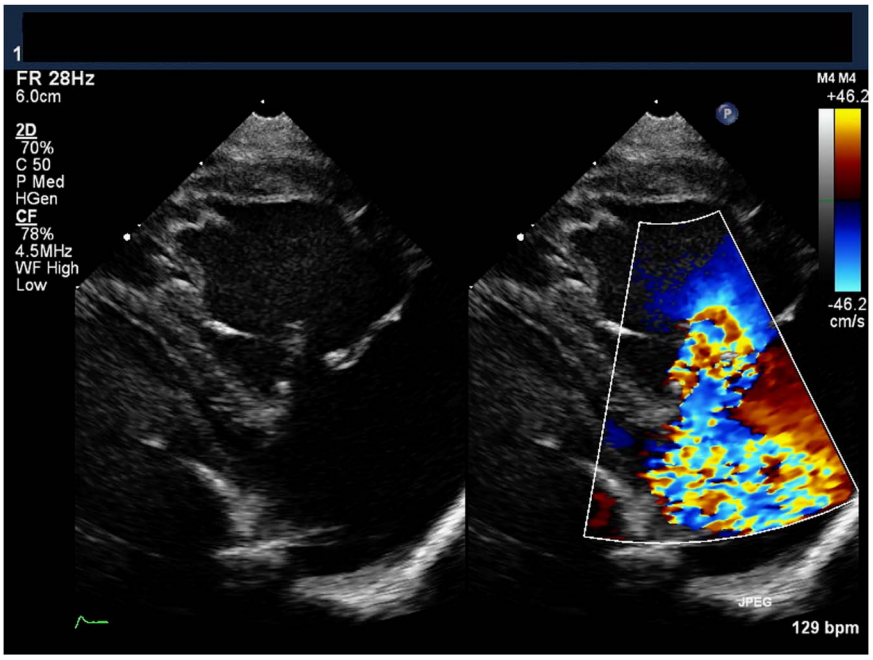

Figure 2.
Neonatal echocardiogram with Doppler showing a flail tricuspid valve and tricuspid regurgitation.

The patient required ventilatory support to maintain saturations above 75%. A strategy of allowing the ductus to become restrictive, followed by pulmonary vasodilator therapy to encourage forward flow across the pulmonary valve was utilised.[4] Cardiac function was initially supported with dobutamine.

Echocardiogram on day 2 and 6 showed progressive improvement of antegrade flow across the pulmonary valve, in keeping with falling vascular resistance, and normal left and right ventricular function. The patent ductus arteriosus had reduced in size, with flow now purely left to right. Ongoing flail anterior leaflet of the

https://doi.org/10.1017/S1047951117000798 Published online by Cambridge University Press

tricuspid valve with associated severe regurgitation was observed. The patient was discharged on day 14 of life, haemodynamically stable, with saturations of 92%. As the patient was clinically stable and growing, surgical opinion and consensus were to delay repair.

At 11 months of age, the patient underwent tricuspid valve repair, patent foramen ovale closure, and atrial reduction without complications. Intraoperatively, tricuspid valve dysplasia with torrential regurgitation and a gigantic right atrium plus significant right ventricular dilatation were noted. Well-developed tricuspid leaflets were observed with complete absence of the suspension apparatus of the anterior and posterior leaflets. Only one identifiable papillary muscle of the septal leaflet was observed. Eight Gore-Tex chordae (Gore Medical, Flagstaff, Arizona, United States of America) were utilised to suspend the leaflets to a prominent ridge on the septum. The annulus was reduced via annuloplasty to a diameter of 19 mm. Postoperative echocardiogram showed mild-to-moderate tricuspid regurgitation, moderately dilated right atrium, and mildly dilated right ventricle with mildly reduced systolic function. Her left ventricular size and function were normal.

Review at 15 months of age revealed oxygen saturations of 100% with echocardiogram findings of mild tricuspid incompetence with low pulmonary artery pressures and normal ventricular size and function (Supplementary video 4).

Discussion

Flail tricuspid valve leaflet secondary to papillary muscle rupture is a rare condition with perinatal mortality up to 80%.[1–3] Early recognition and treatment is imperative for good outcomes.[3] Haemodynamically, the presentation can vary from virtually asymptomatic to extreme cyanosis and cardiac arrest shortly after birth. In cases such as the one described here, presentation is similar to severe Ebstein's Disease, with functional pulmonary atresia from reduced right ventricular ejection across the pulmonary valve and relatively high pulmonary pressures in early life. Ductal patency can lead to a flow-reversal circuit from the duct to the pulmonary artery into the right ventricle, back through the tricuspid valve, and right to left across an atrial septal communication.

Management requires encouragement of forward flow across the pulmonary valve by allowing restriction in left-to-right ductal flow, promoting a reduction in pulmonary vascular resistance, and volume replacement. In some cases, prostaglandins have been used to reduce pulmonary vascular resistance, and extracorporeal membrane oxygenation is required as a bridge to emergency repair of the papillary muscle attachments.

The aetiology of neonatal papillary muscle rupture is often unknown[1,5] and a variety of factors have been weakly implicated, including myocardial ischaemia secondary to hypoxia or premature ductus arteriosus closure, CHD, rhesus isoimmunisation, thromboembolic events, birth trauma, coagulopathy, congenital infective endocarditis, and maternal connective tissue and autoimmune disorders[1,2,6,7].

The presence of maternal anti-SSA antibodies are well known to be associated with congenital complete heart block and occasionally cardiomyopathy. There are cases of endocardial fibroelastosis associated with anti-SSA/SSB in the absence of complete heart block.[8] A fetal immune response involving the papillary muscles could contribute to rupture by an inflammatory process. The presence of maternal antiphospholipid syndrome in this case may represent a causative factor for papillary muscle rupture. A single previous case report described flail tricuspid valve leaflet in a 3-month-old infant with congenital complete heart block and maternal anti-SSA/Ro; however, there were confounding factors of antenatal dexamethasone usage and pulmonary valve stenosis, which may have been contributing factors.[7]

Unlike many previously reported cases of flail tricuspid valve leaflets in neonates, in this case, the time course of the disease process in utero could be observed: echogenic papillary muscles were noted at 21 weeks, and valve patency was confirmed at 27 weeks of gestation. Rupture therefore occurred during the third trimester or the intrapartum period. Echogenic chordae and papillary muscles have been observed in case reports of papillary muscle rupture;[2,3,9] however, the clinical significance of isolated echogenicity, observed in an otherwise structurally normal heart, is not completely understood.

In summary, we describe a case of flail anterior leaflet of the tricuspid valve due to papillary rupture occurring in the peripartum period, presenting in a neonate with a maternal history of anti-SSA antibodies and antiphospholipid syndrome. When the combination of maternal autoantibodies and echogenic papillary muscles is identified in utero, it may be prudent to recommend delivery at a tertiary unit with neonatal intensive care available, as prompt supportive treatment may be necessary and life-saving, as well as good outcome can be anticipated following definitive repair.

Acknowledgements

The authors would like to thank the patient's family for kindly consenting to this brief report. They also like to thank A/Prof Brizard and the Cardiac Surgery Unit at Royal Children's Hospital, Melbourne for their care of the patient and the Children's Cardiac Centre at Princess Margaret Hospital, Perth for their ongoing support.

Financial Support

This research received no specific grant from any funding agency or from commercial or not-for-profit sectors.

Conflicts of Interest

None.

Ethical Standards

The authors assert that all procedures contributing to this study comply with the ethical standards of the relevant national guidelines. Consent was obtained without obligation, for writing and publication.

Supplementary material

To view supplementary material for this article, please visit https://doi.org/10.1017/S1047951117000798

References

1. Yemul MA, Flail A. Anterior tricuspid leaflet secondary to antero-posterior papillary muscle rupture in a neonate. Echocardiography 2014; 31: E151–E155.
2. Loftus PD, Arrington CB, Kaza AK. Neonatal flail tricuspid valve: diagnosis and management. Ann Thorac Surg 2014; 98: 1098–1101.
3. Sachdeva R, Fiser RT, Morrow WR, Cava JR, Ghanayem NS, Jaquiss RDB. Ruptured tricuspid valve papillary muscle: a treatable cause of neonatal cyanosis. Ann Thorac Surg 2007; 83: 680–682.
4. Wald RM, Adatia I, Van Arsdell GS, Hornberger LK. Relation of limiting ductal patency to survival in neonatal Ebstein's anomaly. Am J Cardiol 2005; 96: 851–856.
5. Anagnostopoulos PV, Alphonso N, Nölke L, et al. Neonatal mitral and tricuspid valve repair for in utero papillary muscle rupture. Ann Thorac Surg 2007; 83: 1458–1462.
6. Kaulitz R, Haen S, Sieverding L, Ziemer G. Intrauterine rupture of anterior tricuspid valve papillary muscle: tricuspid valve chordae replacement on the first day of life. J Thorac Cardiovasc Surg 2012; 143: 241–243.
7. Fleming GA, Scholl FG, Kavanaugh-McHugh A, Liske MR. A case of an infant with flail tricuspid valve due to spontaneous papillary muscle rupture: was neonatal lupus the culprit? Pediatr Cardiol 2008; 29: 442–445.
8. Nield LE, Silverman ED, Smallhorn JF, et al. Endocardial fibroelastosis associated with maternal anti-Ro and anti-La antibodies in the absence of atrio-ventricular block. J Am Coll Cardiol 2002; 40: 796–802.
9. Min J, Kim ER, Yang CK, Kim W, Jang WS, Cho S. Successful repair of critical tricuspid regurgitation secondary to a ruptured papillary muscle in a neonate. Korean J Thorac Cardiovasc Surg 2014; 47: 398–401.